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Case Report

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Primary left atrial rhabdomyosarcoma in a 42-year old patient: an unusual cardiac tumor presentation

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ABSTRACT

Left atrial rhabdomyosarcoma is an extremely rare and often overlooked form of primary cardiac malignancy that primarily affects the atria of the heart. The aggressive nature of atrial rhabdomyosarcoma necessitates early recognition, a comprehensive diagnostic workup, and prompt intervention to achieve the best possible outcomes. In this case report, we present the clinical details of a 42-year-old female diagnosed with left atrial rhabdomyosarcoma, who exhibited symptoms of recent pulmonary edema, weight loss, and alternating flushing. Diagnosis, surgical treatment, and the iatrogenic atrial septal defect complication due to interatrial septum invasion were discussed.

Keywords: Primary cardiac tumor, rhabdomyosarcoma, cardiooncology

INTRODUCTION

Atrial rhabdomyosarcoma is an extremely rare and often overlooked form of primary cardiac malignancy that primarily affects the atria of the heart. This neoplasm, characterized by its aggressive nature and predilection for young individuals, poses a distinctive challenge for both clinicians and researchers. Its scarcity and unique clinical presentation have contributed diagnostic challenges, and a dearth of well-documented cases in the medical literature.[1] The management strategy of such tumors is not well defined due to the insufficient data on primary malignant cardiac neoplasms.[1,2] In this case report, we shed light on the clinical, diagnostic, and therapeutic aspects of a case of atrial rhabdomyosarcoma presenting with symptoms of heart failure (HF) due to left atrial (LA) invasion, with the aim of enhancing our understanding of this rare cardiac tumor and its potential implications for patient care.

CASE

A 42-year-old female presented to the cardiology clinic with complaints of exertional dyspnea classified as New York Heart Association class 3, a history of recent pulmonary edema, weight loss and alternating flushing. The patient had no significant previous infections or operations. Her family history of cardiac disease was not significant. Her past medical history was unremarkable, and she did not take any medications, regularly consume alcohol, or use any illicit substances. Upon evaluation, the patient exhibited symptoms of dyspnea and orthopnea.

The patient's vitals showed a blood pressure of 126/87 mmHg, body temperature of 36.2°C, and a heart rate of 96 bpm. Physical exam was notable for inspiratory sounds on the left lower zone and a mild to moderate (3/6) holosystolic murmur at the apex. Additionally, her ECG revealed sinus tachycardia,

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otherwise within normal ranges (QRS interval: 110 ms, QTc interval: 402 ms). The biochemical panel was within normal limits except high sensitive C reactive protein was elevated at 32 mg/L. Transthoracic echocardiography (TTE) demonstrated left ventricular ejection fraction to be 60%, moderate mitral regurgitation, and a suspicious mass in the LA. Subsequently, transesophageal echocardiography (TEE) was performed, and a 24x11 mm non-mobile mass was detected in the posterior wall of the left atrium which elongated through the interatrial septum (IAS) (Figure 1).

The mass was surgically removed by clean dissection along the atrial wall and IAS. Upon surgical inspection, the tumor encountered in the posterior atrial wall surface seemed to be roughened and elongated to the IAS. Tumor infiltration of the IAS was evident. A partial defect in IAS was left as a result of the extensive excision. Due to the high possibility of recurrence and the necessity of serial follow-up, IAS was not closed with a surgical patch. There was no evidence of pericardial or other adjacent structural spread of the mass.

Histological examination of the tumor revealed that the presence of spindle cells with eosinophilic cytoplasm and a fascicular pattern which stained positive for desmin and caldesmon, consistent with rhabdomyosarcoma (Figure 2).

In the postoperative period, a follow-up TTE and TEE did not detect any residual mass lesion in the left atrium (LA). However, a left-to-right shunt was observed from an approximately 10 mm sized defect in the posterior part of the interatrial septum (IAS) (Figure 3). This shunt was determined as restrictive, and close monitoring was recommended. With the oncology consultation, 8 cycles of adjuvant chemotherapy regimen involving vincristine, dactinomycin, cyclophosphamide (VAC) with serial cardiac function monitoring (TTE). Following treatment, the patient did not experience a reoccurrence of dyspnea or other symptoms at 9-month follow up and remains under medical surveillance. Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Figure 1. Transesophageal echocardiography images. Left panel: Two chamber view demonstrated (arrow) hypoechogenic left atrial mass. Right panel: Modified bi-atrial view demonstrated increased thickness and heterogenous appearance of interatrial septum.

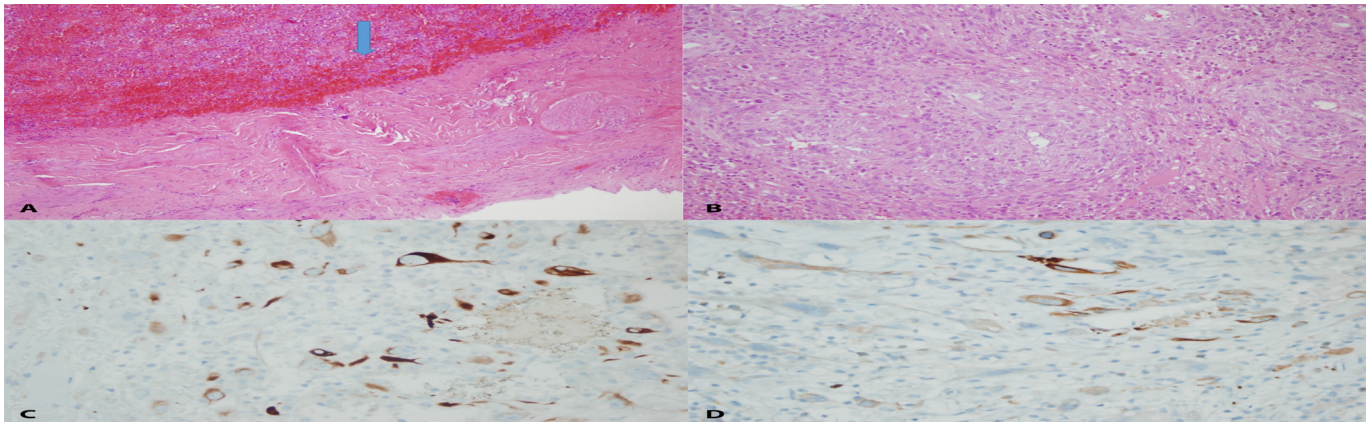

Figure 2. Histological examination of the tumor. A: Blue arrow demonstrated the infiltration of tumor in myocardial wall (H&E, X100 magnification). B: Tumoral cells forming fascicular pattern spindle cells with eosinophilic cytoplasm (H&E, X200 magnification). C: Intracytoplasmic reaction positive for Desmin (IP stain for Desmin, X400). D: Caldesmon positive tumor cells (X400 magnification). Abbreviations: H&E: Hematoxylin & Eosin stain, IP: immunoperoxidase stain.

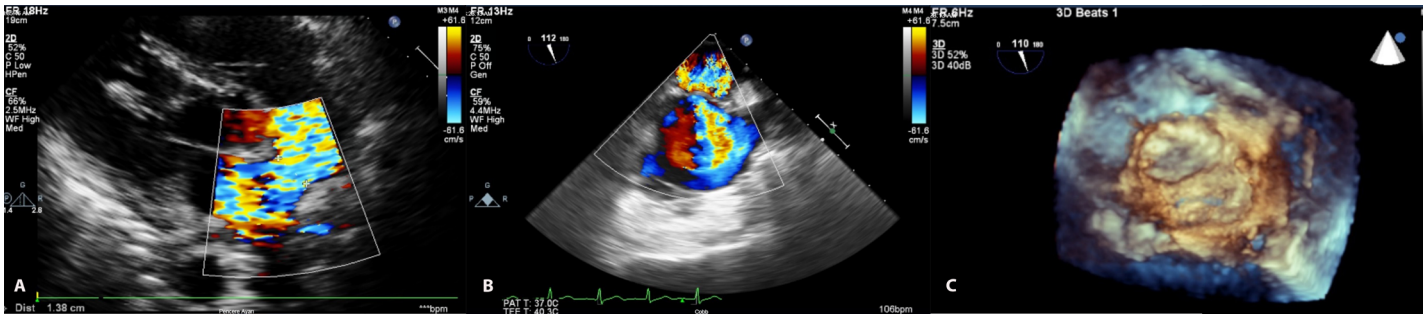

Figure 3. Postoperative TTE and TEE images. A: Modified subcostal plane imaging demonstrated large interatrial defect after surgery. B: Modified bi-caval view demonstrated left to right shunt after surgery. C: 3-Dimensional reconstruction demonstrated a slit-like defect in IAS. Abbreviations: TTE: Transthoracic echocardiography, TEE: Transesophageal echocardiography, IAS: interatrial septum.

DISCUSSION

Cardiac tumors, though relatively rare compared to tumors in other parts of the body, represent a diverse group of neoplasms that can affect various locations while mostly affecting cardiopulmonary functioning.[1,3] While a great portion of them present asymptomatically, we report a case of a patient who experienced HF symptoms as a complication of LA rhabdomyosarcoma with systemic symptoms.

Considering the fact that the heart is one of the rarest locations for tumorigenesis, primary cardiac tumors constitute a minority of all cardiac tumors which incidence was reported as 0.0017% to 0.28%.[1,2,4] Among these primary cardiac tumors, approximately 25% are malignant in nature, with rhabdomyosarcoma being one of the most prevalent malignant examples.[1,2] Malignant neoplasms encompass a range of tumors, including angiosarcomas, rhabdomyosarcomas, fibrosarcomas, and osteosarcomas.[5,6] The behavior of malignant neoplasms within the heart varies based on their specific subtype, histological characteristics, and invasiveness.[6] The determination of the subtype is best achieved through the application of immunohistochemical techniques.[6]

Clinical manifestations and symptoms associated with these cardiac tumors can differ based on their location within the heart, primarily found in the atria and their propensity for growth. They can be originated from various locations within the heart.[1] Although these neoplasms originate intramurally, they can present with complaints of dizziness and chest pain and can lead to arrhythmias and pericardial effusions, which can advance to tamponade, as a result of transmural invasion.[1] Alternatively, they can lead to symptoms similar to those in our patient, stemming from the obstruction of cardiac cavities, including the right and left atrium (RA/LA), inferior vena cava, and others in relation to the neighboring structure they are mainly affecting.[3,7,8] In cases where a tumor invades the right side of the heart, the presentation can often resemble that of tricuspid or pulmonary valve diseases, with signs of right heart failure.[1,7] Tumor metastases tend to be detected in various organs, including the lung, liver, thoracic lymph nodes, and pancreas.[2,3,8] Additionally, on rare occasions, cardiac rhabdomyosarcoma may give rise to cerebral embolisms.[2]

Atrial rhabdomyosarcoma may manifest with systemic symptoms such as weight loss, night sweats, and fever.[2,7,8] The presence of B symptoms and flushing can be the primary complaints of the patients. Recognizing these systemic symptoms is crucial for early detection and intervention, as they can provide valuable clues in the comprehensive assessment of patients with atrial rhabdomyosarcoma. Physicians should keep in mind about possible diagnosis of atrial rhabdomyosarcoma as a differential in patients presenting with similar complaints.

Various imaging studies, such as TTE, TEE, computed tomography, coronary angiography, and magnetic resonance imaging, play a pivotal role in both diagnosis and guiding surgical interventions.[7,8] Serial TTE and TEE should be considered throughout the presentation. However, definitive diagnosis relies heavily on tissue biopsy and subsequent histopathological analysis.

Classical treatment modalities for managing these cardiac tumors typically involve a combination of surgical resection, chemotherapy, and radiotherapy. In our patient's case, surgical resection, and chemotherapy of were employed, and a restrictive IAS defect was identified as a complication following tumor excision surgery. Through this case, we aimed to underline the importance of early recognition and multidisciplinary management in improving the prognosis of this intriguing and uncommon malignancy.

CONCLUSION

The aggressive nature of atrial rhabdomyosarcoma necessitates early recognition, a comprehensive diagnostic workup, and prompt intervention to achieve the best possible outcomes. In the absence of standardized treatment protocols for this rare malignancy, the establishment of an individualized, patient-specific therapeutic plan is paramount. This case report underscores the need to consider atrial rhabdomyosarcoma as part of the differential in cardiopulmonary disease, especially those in young, otherwise healthy individuals, as the limited data available hinders our understanding of this rare condition. Further investigations and clinical experiences are warranted to advance our knowledge and enhance the care provided to those affected by this exceptionally rare disease.

ETHICAL DECLARATIONS

Informed Consent

The patients signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

REFERENCES:

1. Tyebally S, Chen D, Bhattacharyya S, et al. Cardiac tumors: JACC CardioOncology state-of-the-art review. *JACC CardioOncol.* 2020;(2):293-311.
2. Chlumsky J, Hola D, Hlavacek K, et al. Cardiac rhabdomyosarcoma. *Exp Clin Cardiol.* 2001;(2): 114.
3. Rasekhi RT, Hosseini N, Babapoor-Farrokhran S, et al. A rare case of primary adult cardiac rhabdomyosarcoma with lower extremity metastasis. *Radiol Case Rep.* 2023;(18):1666-1670.
4. Castorino F, Masiello P, Quattrocchi E, et al. Primary cardiac rhabdomyosarcoma of the left atrium: an unusual presentation. *Tex Heart Inst J.* 2000;(27):206-208.
5. Dirican A, Kucukzeybek Y, Erten C, et al. Cardiac rhabdomyosarcoma of the left atrium. *Contemp Oncol (Pozn).* 2014;(18):73-75.
6. Chen J, Liu X, Lan J, et al. Rhabdomyosarcoma in adults: case series and literature review. *Int J Womens Health.* 2022;(14):405-414.
7. Uchida T, Kuroda Y, Sadahiro M. Primary biatrial cardiac rhabdomyosarcoma. *Braz J Cardiovasc Surg.* 2020;(35):399-401.
8. Ma J, Sun JP, Chen M, et al. Left atrial rhabdomyosarcoma. *Circulation.* 2014;(129):e503-e505.